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MALIGNANT SMALL CELL (ASKIN) TUMOR OF THE THORACOPULMONARY REGION ORIGINATING FROM THE PHRENIC NERVE

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Malignant small cell tumors of the thoracopulmonary region, first described by Askin and colleagues,[1] are rare, malignant neoplasms belonging to the family of peripheral primitive neuroectodermal tumors. We report a case of malignant small cell tumor originating from the left phrenic nerve in a 31-year-old woman with a hemothorax.

Clinical summary. In 1983, when she was 16 years old, the patient underwent a left thoracotomy and pleurectomy for spontaneous pneumothorax after failure of conservative treatment. Although she had no recurrence of pneumothorax, a residual anterior basal pleural air space was documented on subsequent follow-up radiographs and on a computed tomographic scan obtained in 1987.

She was recently referred to us for a left basal pleural effusion accidentally detected during a gynecologic ultrasound examination. A subsequent chest x-ray film showed an opacification of the old basal pleural cavity. Magnetic resonance imaging of the chest displayed a heterogeneous round lesion 7 cm in diameter within a high intensity signal from a left anterior basal effusion (Fig 1).

The woman was completely free of symptoms, and physical examination was unrewarding. A basal thoracentesis yielded hemorrhagic fluid that was negative for bacteria, mycobacteria, and atypical cells. A fiberoptic bronchoscope showed a completely normal airways system.

A biopsy specimen of the lesion was obtained by thoracoscopy in the basal cavity. Histologic examination revealed a round small cell tumor with a high proliferation rate. Thus the patient underwent a left thoracotomy and excision of a round, bosselated, whitish gray, partly cystic mass that was located in the anterior cardiophrenic angle, with the base on the central tendon of the diaphragm. The tumor appeared to arise from the sternal branch of the phrenic nerve at its origin, which was resected along with the mass. The latter was capsulated and easily enucleated from the surrounding structures. The postoperative course was uneventful and the patient was discharged on postoperative day 6.

At 1 year's follow-up, the patient had a local recurrence but was free of distant metastases.

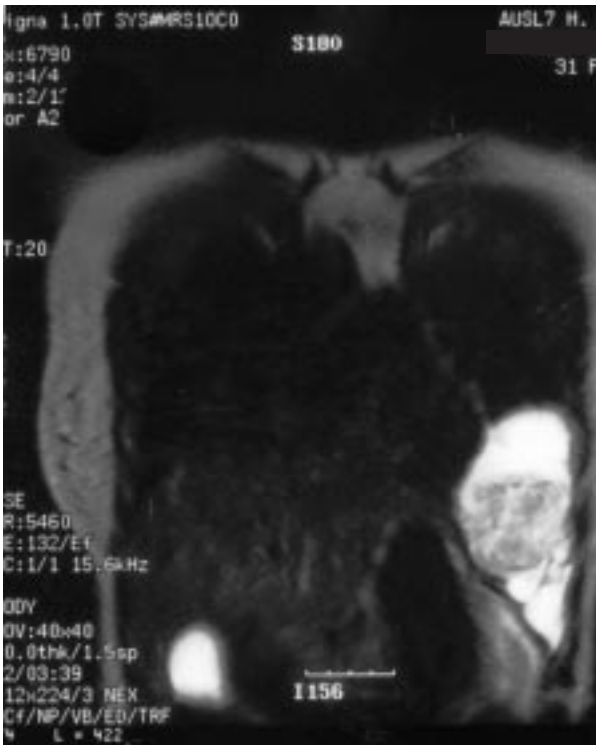

Fig 1. Magnetic resonance T2-weighted coronal image of the chest showing a heterogeneous mass within a high-intensity signal hemorrhagic effusion in the left basal pleural space.

Discussion. Malignant small cell tumors of the thoracopulmonary region most likely originate from the intercostal nerves and may invovle the ribs, the soft tissue of the chest wall, and the lung, which sometimes can be the primitive site, but they have never been described associated with the phrenic nerve.

A differential diagnosis has to be made with other small cell tumors such as Ewing's sarcoma, rhabdomyosarcoma, malignant lymphoma, and neuroblastoma.

Histologic examination showed small round malignant cells with scant eosinophilic cytoplasm, high nuclear/cytoplasmic ratio, small single nucleoli, and a high mitotic rate. The cells often had an epithelioid aspect and were arranged in solid aggregates interspersed with mixoid tissue and large necrotic and hemorrhagic areas. Occasionally rosette-like structures were evident, but the lack of typical Homer-Wright rosettes excluded neuroblastoma.[2]

By immunohistochemistry, the tumor was strongly positive for neuron-specific enolase, which is a specific marker for neural elements,[3,4] even though Ewing's sarcoma and rhab-

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domyosarcoma may display neuron-specific enolase positivity[3]; vimentin, commonly positive in neuroectodermal tumors[4]; and for HBA 71. Immunoreactivity for leukocyte common antigen (CD45), chromogranin, synaptophisin, S-100 protein, myeloperoxidase, neurofilaments, and glial fibrillary acid protein was negative. Although these latter antibodies showed variable results in other reports,[4] the absence of reaction for chromogranin and the strong immunoreactivity for HBA 71 differentiate primitive neuroectodermal tumor from neuroblastoma.[5]

Ultrastructural studies revealed dense core neurosecretory granules, short cytoplasmic processes that were highly suggestive of neural differentiation, filamentous cytoskeleton, and prominent Golgi bodies. These features of primitive neural lymphoma[3] and the absence of gangliocytic and Schwann cell differentiation distinguish this tumor from neuroblastoma.[5] Moreover, the lack of cross striation and the prominent Golgi bodies excluded rhabdomyosarcoma.[2,3]

Magnetic resonance images showed in our case a heterogeneous pattern suggesting hemorrhage and necrotic areas, which are characteristic of malignant small cell tumors.

A possible relationship between the old pleurectomy operation and the subsequent residual basal air space and the occurrence of this tumor from the phrenic nerve in that same area is tempting but merely speculative (ie, mechanical factors, chronic inflammatory stimuli).

Although multimodality treatment is recommended, the prognosis for this type of neoplasms is dismal because of the high likelihood of local recurrence and distant metastases.

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